An Unusual Case of Reversible Empty Sella

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Abstract: Context: An empty sella is a relatively common condition, often being an incidental finding at MRI or CT scan. It can develop because of the intrasellar herniation of Cerebro-spinal Fluid (CSF) and arachnoid membrane through an absent or rudimentary diaphragm sellae in concomitance of a sudden and even transient increment of intracranial pressure, leading to a picture in which the pituitary is flattened along the floor of the sella.

Case Description: A young female with headache, nausea, dizziness, diplopia and visual impairment showed an empty sella on MRI and increased CSF pressure at the lumbar puncture. After an initial improvement, there was a progressive worsening of the headache, especially in orthostatic position, with transient relief after bed rest and hydration. At MRI the empty sella was no longer evident, cerebellar tonsils were displaced in the occipital foramen and there was an impregnation of the meninges after contrast medium, a picture of CSF hypotension, probably due to the previously performed lumbar puncture causing a fistula with leak of CSF and consequent disappearance of the empty sella. The patient gradually improved after being submitted to epidural blood patch.

Conclusions: The case here reported demonstrates that an empty sella can be a reversible condition in rare cases. Its disappearance can be due to the reduction in intracranial pressure caused by the lumbar puncture itself. The changes in the characteristics of the headache, in particular its worsening in the orthostatic position, should lead to the suspicion of CSF leak through a fistula and consequent intracranial hypotension, a dangerous and sometimes life-threatening condition.

Keywords: Cerebrospinal fluid hypotension, empty sella, intracranial hypertension, meningeal leak.

CLINICAL CASE

A 21 years old female complaining about the recent onset of a progressive worsening headache followed by nausea, objective dizziness, photophobia, diplopia, visual impairment, fatigue, anxiety and panic attacks was initially evaluated for the possible presence of a cerebral mass with intracranial hypertension.

The past clinical history was that of a healthy young woman, with irregular menses and with no previous episodes of headache or visual problems nor habitual use of drugs. The neurological symptoms had begun about 48 hours before.

She was overweight (weight 73 Kg, height 163 cm, BMI 27.5 Kg/m2, waist circumference 86 cm), with normal distribution of body hair, and normal cardio-pulmonary and abdominal findings.

There were no focal neurological signs at clinical examination. The electroencephalogram (EEG) was normal.

She was initially evaluated by an ophthalmologist that, however, despite the referred symptoms, didn’t find a papilledema at fundoscopic examination, nor altered visual field or pathological response to retinal evoked potentials.

There was an empty sella with a flattened pituitary gland along the floor of the sella and normal ventricular size at Magnetic Resonance Imaging (MRI) (Fig. 1). Then, there was no evidence of hydrocephalus, mass or vascular lesions at MRI.

A lumbar puncture showed an increased Cerebro-Spinal Fluid (CSF) opening pressure (250 mmH2O), with a normal composition of the CSF itself.

Hormonal assessment showed normal FSH, LH, 17-beta estradiol, FT4, FT3, ACTH and Cortisol (even after ACTH 1 mcg iv stimulation). PRL and TSH were slightly augmented whereas IGF1 was slightly reduced (157 ng/ml) and the GH response to GHRH+Arginine was blunted (peak value of 5.4 ng/ml at 120').

The intracranial hypertension was possibly referred as the consequence of the use of an estro-progestinic preparation (etinilestradiol 0.05 mg + levonorgestrel 0.25 mg) same days before. Clinical conditions slightly improved in the following days and the patient was discharged with 12.5 mcg
of levo-thyroxine to treat subclinical hypothyroidism and Venlafaxine 75 mg/day for anxiety and panic attacks.

She came back after three months, however, complaining about the progressive worsening of the headache and the onset of cervical pain, with scarce response to analgesics and transient relief after prolonged bed rest in clinostatic position and hydration with normal saline. At MRI (Fig. 2) the picture had been completely changed: the empty sella was no longer evident as the pituitary gland had reassumed its normal position inside the sella. The peduncle was dislocated on the right with no clear evidence of a pituitary adenoma. The cerebellar tonsils were displaced in the occipital foramen, reaching the C1-C2 level.

The quality of life, and especially social life were deeply impaired because the patient was obliged to maintain a supine position for most of the time, in order to obtain a transient relief of the headache. The mood was depressed and the patient was very worried about her clinical condition. There was no improvement of clinical status over the time.

Another MRI 3 months later (Fig. 3) showed also an enhancement of the dura mater and of the meninges of the internal acoustic meatum after contrast medium administration and dilatation of the suprachiasmatic cistern. The picture was that of a CSF hypotension probably due to the previously performed lumbar puncture causing a meningeal leak of CSF.

The patient was submitted to epidural blood patch at lumbar level. The clinical as well as the MRI picture gradually improved during the following months, with the gradual disappearance of the enhancement of the meninges. The hormonal assessment showed normal values and a normalized GH-IGF-1 axes and TSH. The patient is still on follow-up.

**COMMENT**

An empty sella is a relatively common condition, often being an incidental finding at MRI or CT scan. It can develop because of the intrasellar herniation of CSF and arachnoid membrane through an absent or rudimentary diaphragm sellae in concomitance of a sudden and even transient increment of intracranial pressure, leading to a picture in which the pituitary is flattened along the floor of the sella [1].

One of the causes of empty sella is the so-called Pseudotumor Cerebri or idiopathic (benign) intracranial hypertension, a rare condition of unknown causes characterized by increased intracranial pressure, headache and papilledema with associated visual impairment or loss, with no focal neurological signs, most often seen in obese women and occasionally during pregnancy [2]. Other causes of intracranial hypertension, such as obstructive hydrocephalus, tumors, meningitis and dural sinus thrombosis should be ruled out. Opening lumbar CSF pressure is augmented (>250 mmH2O), whereas CSF composition is normal [3, 4]. Besides obesity, this condition can be associated with irregular menses, Polycystic Ovary Syndrome (PCOS), steroid withdrawal, hypothyroidism, hypo- and hyperparathyroidism, Guillain-Barré syndrome, drugs such as vitamin A, tetracycline, lithium and estro-progestin compounds [5, 6].

The possible mechanisms involved in the development of the increased intracranial CSF pressure are both vasogenic extracellular brain edema and a low conductance of CSF outflow at the level of arachnoid villi, with the subsequent compression of intracranial venous sinuses by the increased intracranial pressure that, in turn, further reduces the CSF flow across the arachnoid villi [6]. Both endogenous and exogenous estrogens can promote or worsen idiopathic intracranial hypertension, especially in conditions characterized by trombophilia and hypofibrinolysis (obesity, PCOS, pregnancy) [7].
Glucocorticoids, acetazolamide, and diuretics, as well as serial lumbar punctures have been used to lower the intracranial pressure, especially in case of visual impairment. The reduction in CSF pressure can ameliorate the headache and visual impairment and even cause a regression of the empty sella [8]. Lumbar puncture, however, can be followed by headache because of the transient CSF hypotension, with a rapid regression after adequate bed rest. Sometimes, however, a persistent leak of CSF can be due to the formation of a fistula. In this case the headache tend to persist and characteristically it worsens in orthostatic position and slightly improves after prolonged bed rest and hydration. It can severely impair the quality of life. A marked reduction in CSF pressure can lead to cerebellar tonsils herniation, with possible compression of the brain stem, a very dangerous and sometimes life-threatening condition. At MRI there are a characteristic impregnation of the meninges after contrast medium administration.

A possible therapy can be represented by epidural blood patch. The volume of the autologous blood injected in the epidural space directly increases CSF pressure, thus reducing traction on brain and meningeal structures, leading to the relief of symptoms; furthermore, the blood creates a clot particularly adherent to the dura mater, patching the hole in the dura and closing the fistula, thus avoiding further CSF leak. The epidural blood patch can restore a normal intracranial pressure, ameliorating the clinical picture [9].

The case here reported is an unusual case of reversible empty sella. The herniation of subarachnoid spaces and CSF into the sella was due to a transient increment of intracranial pressure possibly caused by the ingestion of an estro-progestin association. The following re-expansion of the pituitary gland and the disappearance of the empty sella, associated with a deep change in the characteristics of the headache (in particular the postural response) were both caused by the reduction in intracranial pressure (and then CSF hypotension) due to the leakage of CSF through a fistula in the dura mater at lumbar level caused by the previously performed lumbar puncture.

CONFLICT OF INTEREST
The authors confirm that this article content has no conflict of interest.

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REFERENCES